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Carotico-cavernous fistula: An educational case

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ABSTRACT

INTRODUCTION: We present a case of direct carotico-cavernous fistula (CCF) and its successful treatment.
PRESENTATION OF CASE: A 55-year-old male presented with left eye discomfort, diplopia and pulsatile tinnitus. He had a recent history of head injury. Examination showed proptosis, chemosis, orbital bruit, and abducens nerve palsy. Digital subtraction angiography confirmed CCF, which was managed endovascularly. The patient recovered fully after treatment.
DISCUSSION: CCF has a variety of presenting clinical signs, imaging manifestations, and treatment options available.
CONCLUSION: CCF is a rare and dangerous neurological disorder that should be promptly diagnosed and treated.

1. Introduction

Carotico-cavernous fistula (CCF) is a rare sight- and life-threatening disorder that arises due to an abnormal connection between the carotid artery and the cavernous sinus. We present a case of CCF in which the diagnosis was missed at initial presentation, but was subsequently successfully treated with good outcome.

2. Presentation of case

A 55-year-old male presented with a 1-week history of left eye discomfort, double vision and pulsatile tinnitus. He had a recent history of head injury. CT brain showed only a dilated superior ophthalmic vein (Fig. 1). This was missed and he was discharged home, representing a week later with symptom progression. Examination of the left eye showed proptosis, chemosis, orbital bruit, and sixth cranial nerve palsy. Visual acuity was 6/6 in both eyes, and there were no other neurological findings.

Cerebral angiography confirmed the presence of a high flow fistulous connection between the internal carotid artery and the cavernous sinus by demonstrating rapid filling of the cavernous sinus following internal carotid arterial injection (Fig. 2). Treatment was effected successfully by transarterial embolization using several platinum coils (Fig. 3). All symptoms fully resolved following treatment.

3. Discussion

Dilatation of superior ophthalmic vein (SOV) was initially missed in this patient. There are a variety of causes of this abnormality, ophthalmic Graves' disease, Tolosa-Hunt syndrome, inflammation at the apex of the orbit, ocular vascular deformity, cavernous sinus tumour, and thrombosis of the cavernous sinus. However, it has been suggested that nearly 80% of cases of superior ophthalmic vein dilatation, as found on imaging, were the result of carotico-cavernous fistula, whereas only 12% were the result of Graves' disease, and many fewer were the result of other aetiologies.[1] Clinical examination, and patient history will often point to the correct diagnosis.

CCF arises due to the formation of an abnormal communication between the carotid artery and the cavernous sinus. This may be traumatic or spontaneous. Direct CCF (type A) is most often traumatic, as in this case, and is caused by rupture of the ICA in its intracavernous course. Indirect CCF (types B–D) results from the spontaneous rupture of dural branches of the intracavernous arteries usually causing low-flow fistulae, and is often associated with hypertension, atherosclerosis, or collagen disorders. The classification of CCF is shown in Table 1.

Common clinical findings[1] in cases of direct CCF develop suddenly, and include: orbital bruit (80%), proptosis (72%), chemosis (55%), abducens nerve palsy (49%), conjunctival injection (44%), other rarer findings are legion. The risk of permanent neurological disability is lower in younger patients; the overall risk is 4%.[2]

Angiography gives the definitive diagnosis of carotico-cavernous fistula. Specific signs on imaging can be demonstrated by various modalities[3] including colour Doppler ultrasonography,

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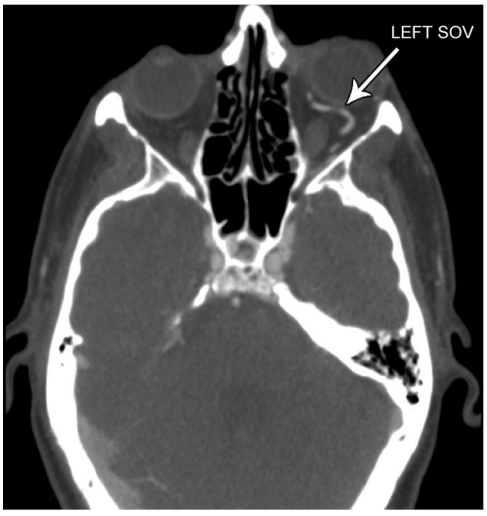

Fig. 1. CT brain showing dilated left superior ophthalmic vein.

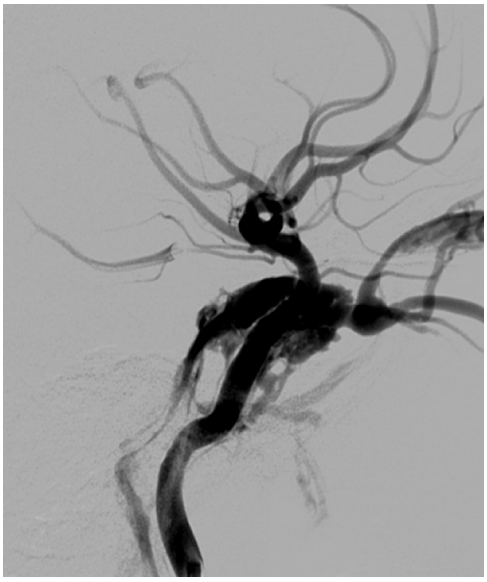

Fig. 2. Direct left internal carotid artery injection, showing early filling of an enlarged cavernous sinus in the arterial phase (cerebral angiography, AP projection).

CT angiography, and MR angiography, however, the gold-standard is DSA due to its superior capability to accurately localize lesions for endovascular management.

Definitive management of obliterating the fistulous connection is most often achieved by an endovascular approach. Prompt treatment is required in cases of direct CCF with progressive symptoms,

Table 1
Classification of carotico-cavernous fistula.

Category	Definition
Type A	Direct high-flow fistulae that are the result of a tear between in the ICA during its intracavernous course; usually traumatic (AI) or due to a ruptured aneurysm (AII).
Type B	These are dural shunts between meningeal branches of the ICA and the cavernous sinus, they are usually spontaneous.
Type C	These are dural shunts between meningeal branches of the external carotid artery (ECA) and the cavernous sinus, they are usually spontaneous.
Type D	These are dural shunts between meningeal branches of both the ICA and the ECA and the cavernous sinus, also spontaneous in origin.

Fig. 3. Post coil embolization showing complete obliteration of the CCF (cerebral angiography, lateral projection).

since visual loss occurs in nearly 90% of untreated patients.[4] Conservative treatment is indicated for small, asymptomatic, or stable fistulae, since these may close spontaneously.

Endovascular treatment is usually effected using the intra-arterial route via the femoral artery. Embolization is most often carried out using detachable balloons, though platinum coils (as used in this case, Fig. 3), liquid embolizing agents, stents or a combination may be used.[5]

Should an endovascular approach be inappropriate, or ineffective, surgical management is an option. Direct repair of the fistula within the cavernous sinus is possible.[6] Trapping of the ICA involves ligation of the supraclinoid carotid and ophthalmic arteries intracranially, followed by ligation of the ICA in the neck. This approach is contraindicated should the collateral blood supply be inadequate.

4. Conclusion

CCF is a rare and dangerous neurological condition that should be promptly diagnosed and treated.

Conflict of interest

The authors report no declarations of interest.

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Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Contributors

Sean Martin has written, reviewed and submitted the article; Mario Teo critically reviewed the article; Jo Bhattacharya critically reviewed article, performed the procedure and obtained images;

Likhith Alakandy critically reviewed article, senior consultant in charge of case.

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